

Behcet's Disease (BD) in Iran; Epidemiology and Clinical Protests

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Behcet's disease (BD), a multi-organ disorder which is classified as vasculitis disease and able to affect all types of vessels, characterized by frequent relapse and remission, and mainly affects the countries of the Silk Road.

The epidemiology and Clinical protests of BD is vary from region to region. On the other side, the image of disease and epidemiology in a region has been changed over the years. Iran is one of the countries that this disease has a widespread and their patients have been registered since 1975 at the Shariati Hospital, Behcet's disease Unit, under the direction of Professor F. Davatchi, and since become 7630 patients.

The editorial will explain the epidemiology and Clinical protests of the disease in Iran.

Epidemiology and demography of BD, by 2010, 6500 patients had been registered in Iran and the prevalence of disease 80 per 100,000, and the mean age of onset disease was 26 years, and in men it was 54.9% and in female 45.1%, and the male to female ratio was 1.22%. In fact, the disease is more common in men.

In terms of Clinical protests, they are divided into two major and minor categories:

Major Clinical protests include oral Aphthosis, genital Aphthosis, skin Lesion including erythema nodosum and Pseudo-folliculitis, and ocular involvement including anterior uveitis and posterior uveitis and retinal vasculitis.

Oral aphthosis, ranging from round to oval shape which is single or multiple numbers, relapsing, with a white to yellow necrotic center Surrounded with an erythematous margin varying from a few millimeters to several centimeters, and the duration of recovery from several days to several weeks but usually the duration is around two weeks. The lesions are fully recovered but Genital and giant oral Aphthosis can leave scars.

The oral aphthosis is the most common Clinical protests of BD and is seen in 97.3% of patients in Iran.

The genital aphthosis are similar to mouth aphthosis but larger and deeper, with a longer recovery period and a higher probability of leaving scar with less prevalence and less relapse, which is seen in 64.6% of Iranian patients.

Skin manifestations is different in Behcet's disease and include below items:

Pseudo-folliculitis, erythema nodosum, skin aphthosis, and pathergy phenomenon.

Other lesions can be seen like pyoderma gangrenosum, erythematous induratum like lesions, suppurative panniculitis.

Pseudo-folliculitis is seen as a dome-shaped pustule with the base of erythema and edematous and mainly seen in the upper and lower limbs which is seen in 64.9% of patients in Iran.

Erythema nodosum in BD is similar to other disorders but with more edema and erythema, and mainly in the lower extremities and anterior legs and seen in 22.2% of patients in Iran.

Skin aphthosis is similar to genital aphthosis seen mainly in the area between the breasts and also could be seen inguinal and buttock region, axillary, sub-mammary space, interdigital region, trunk.

The pathergy test is a vasculitis response to needle trauma and is specified to Behcet's disease.

The Eye lesions is a major manifestations of the disease and is the most important cause of morbidity and retinal vasculitis is very dangerous and very resistant to treatment in Behcet's disease. Uveitis also seen as anterior and posterior. Here, also we have relapse and silence similar to other lesions of BD, but the difference is that in eye lesions, there are a new relapse before the complete recovery of last attack made the complication of accumulation, and in the long time it cause, an important complication which are reduction of visual acuity and blindness.

Eye involvement seen in 56.8% of Iranian patients. Anterior uveitis (AU) and posterior uveitis (PU) and retinal vasculitis (RV) are seen 41%, 44.4%, 30.5%, respectively in the patients.

Minor manifestations of the disease include, joint involvement, neurological involvement, vascular involvement, gastrointestinal involvement and respiratory and cardiovascular system involvement and epididymitis.

Joint manifestations are in different range of arthralgia to polyarthritis and oligoarthritis, and monoarthritis and ankylosing spondylitis and seen in 34.7% of Iranian patients.

Arthralgia, monoarthritis, and oligoarthritis and ankylosing spondylitis are found in 17.6%, 7.2%, 16.8%, and 2% of Iranian patients respectively.

Neuro Behcet's Disease (NBD) is a neurological manifestation, which is very rare, but with bad omen morbidity and mortality. Its manifestations include central and peripheral nervous system involvement. Central nervous system involvement is in the form of parenchymal and non-parenchymal involvement. Parenchymal involvement is with brainstem involvement, hemispheric manifestations, spinal cord manifestations, and Meningoencephalitis. Non parenchymal lesions is with dural sinus thrombosis, arterial occlusion, and aneurysms which are seen in 3.7% of Iranian patient.

Central nervous system involvement is founded in 3% and peripheral in 0.2% of patients.

Gastrointestinal involvement is also very rare but more common than neurological manifestations, as Aphthosis in the entire gastrointestinal tract and with various symptoms of abdominal pain, chronic diarrhea, and gastrointestinal bleeding and perforation which is seen at 7.6% In Iranian patients.

The vascular involvement of this disease is very important, it can be involved in any vessel from capillary and vein and even to the large vessels. Venous involvement is included as superficial phlebitis to deep vein thrombosis and major vein thrombosis (vena cava). Arterial manifestations of disease are in form of aneurysms and thrombosis.

In Iran, superficial phlebitis is 2.2% and deep vein thrombosis is 5.7% and large venous thrombosis is 1.1%. Aneurysms and arterial thrombosis are reported in 0.5% and 0.154% of Iranian patients respectively.

Epididymitis is seen in 4.7% of patients (men).

Cardiac manifestations (0.6%) and pulmonary (0.34%) are very rare symptoms of illness in Iranian patients.

Since BD is a multi-organ disease and with different manifestations in different organs and sometimes it is difficult to diagnose and knowing the epidemiology of disease and clinical manifestations helps to better diagnosis and treatment.

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